

Giant Mature Adrenal Cystic Teratoma in an Infant

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Case report

ABSTRACT

Introduction: Teratomas are derived from embryonic tissues that are typically found in the gonadal and sacrococcygeal regions of adults and children. Primary teratomas in the retroperitoneum are very rare in infant

and primary adrenal teratomas are extremely rare. Early diagnosis and surgical resection are important for effective treatment.

Case report: We report here the case of a histologically unusual adrenal teratoma detected on computed tomography during the workup of abdominal distension 3-month-old male

infant. The evaluation and treatment of this condition and a review of the literature are included in this paper.

Keywords: Adrenal gland, Cystic masses, Infant, Teratomasurgery, Tomography, X-Ray, Computed Diagnosis.

1. INTRODUCTION

Incidence of germ cell tumors has been estimated at about 0.9/100,000 population and of these, teratoma has been reported as the leading fetal and neonatal neoplasm (1, 2). Primary teratomas in the retroperitoneum are very rare in infant and primary adrenal teratomas are extremely rare (3). Most teratomas in this region are secondary to germ cell tumors of the testicles or ovaries. Specifically, in male patients, retroperitoneal germ cell tumors are more likely to have metastasized from the testes than to present as primary tumors (2). We describe a case of mature cystic teratoma that was clinically suggestive of an adrenal tumor. We report the case of a primary adrenal mature teratoma presenting in an infant who was treated at our hospital.

No other physical findings were observed. An ultrasound examination showed an 83 by 80 mm mixed cystic, solid lesion, located in the retroperitoneum, with superior and inferior displacements of a normal appearing spleen, left kidney and liver respectively. CT scan showed a large, cystic, heterogeneous, left sided, retroperitoneal mass extending across the midline (Figure 2). Serum alphafetoprotein level (AFP) was within high normal range for a newborn at 46 nanograms/milliliter (ng/ml). Upon surgical exploration a large, freely mobile, tumor originating from the left adrenal gland was identified. Dissection was carried out to release adhesions between the colon and the anterior surface of the mass and after carefully dissecting the tumor from

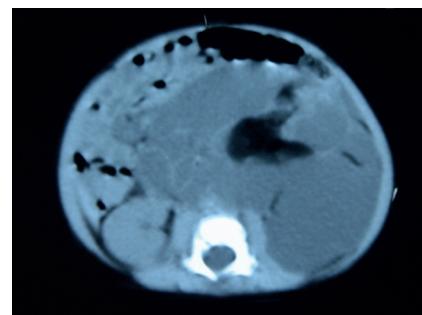


Figure 2. CT scan showed a large, cystic, heterogeneous mass extending across the midline

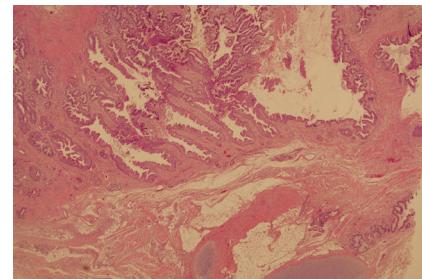


Figure 3. Smooth muscle, cartilage, fat tissue were mixed with mature and immature glial tissue. Arrow shows a glandular structures wall (Hematoxylin-eosin stain, original magnification, X 100).



Figure 1. Abdominal distension in an infant.

the left kidney, abdominal aorta, vena cava inferior, left renal vein, left liver lobe 14 by 10 by 8 cm cystic mass with minimal solid components was removed from the abdominal cavity. Histology showed a solid benign mass with cysts lined by enteric squamous tissue. Smooth muscle, carti-

lage, fat tissue were mixed with mature and immature glial tissue, and glandular structures with dystrophic calcification were present. We found that adrenal gland around of teratoma. This was consistent with a mature teratoma (Figure 3).

3. DISCUSSION

Teratoma is a germ cell tumor derived from totipotential cells, which comprise several parenchymal cell types originating from, also called cystic teratomas, are composed of 3 germ layers (ectoderm, mesoderm, and endoderm) (4). According to various degree of maturation, tumors are classified as mature, immature and malignant teratomas. Most mature teratomas are benign and cystic, which are classically called dermoid cysts. The cyst, which is mainly composed of squamous epithelium, fat, tooth and hair, is a benign neoplasm with malignant potential. Most teratomas are found in the gonads. Nevertheless, many extragonadal sites have been reported, including the mediastinum, retroperitoneum, cranium, sacrococcygeal region, the large bowel, and even the tongue (5, 6, 7).

Most common sites of teratomas in neonates are the sacrococcygeal and presacral regions. In general teratomas that occur in infancy and early childhood are usually extragonadal, whereas those found in older children are more commonly located in the gonads. Retroperitoneal teratomas are extremely rare. There are individual case reports of ectopic tissues located in the adrenal region, which include bronchogenic, nephrogenic, and thyroid tissues, but they lack the range of somatic tissue types associated with a teratoma.

Adrenal teratomas have no specific clinical manifestations. They are often found on ultrasonography. However, abdominal distension, abdominal pain, low back pain, or, even, intestinal obstruction caused by compression of the neoplasm can occur in one half of patients.

The diagnosis of adrenal teratoma relies predominantly on an imaging examination because the findings from laboratory examinations will often be normal. The differential diagnosis of retroperitoneal teratomas

include ovarian tumors, renal cysts, adrenal tumors, retroperitoneal fibromas, Wilms' tumor, sarcomas, hemangiomas, neonatal cystic neuroblastoma, xantogranuloma, congenital mesoblastic nephroma, enlarged lymph nodes and perirenal abscess (8, 9, 10, 11). Plain abdominal film shows a calcification. Sonography can identify the cystic, solid or complex components of the tumor (12). CT defining the teratoma extent to the surrounding organs and in evaluating the cyst wall. 4 Magnetic resonance imaging is better than sonography and CT to demonstrate the anatomical relationship (13). A post-operative pathologic examination has often been required for a definitive diagnosis.

Surgical complete resection and close follow-up should be recommended therapy for mature teratomas and is required for definitive diagnosis (14, 15). However, in the case of an immature teratoma, adjuvant therapy, such as chemotherapy, radiotherapy, or concurrent chemo-radiotherapy, will also be necessary, provided that the primary tumor has been completely resected (16).

The prognosis is excellent for benign retroperitoneal teratoma if complete resection can be accomplished. Usually asymptomatic, large neoplasms can cause abdominal distension. Preoperatively, the diagnosis can be established by its characteristic appearance on radiologic evaluation and biocemical study. The definitive treatment for these neoplasms is surgical resection.

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